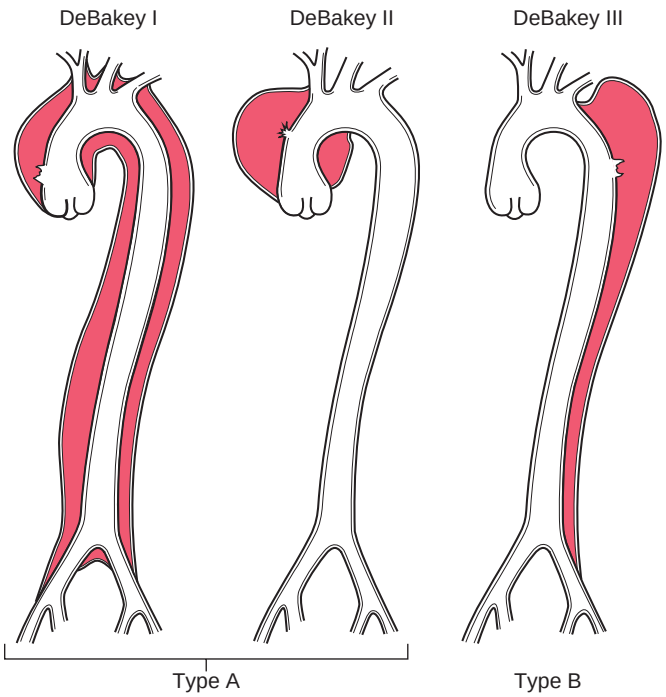

Figure 11-22 Classification of dissections. Type A (proximal) involves the ascending aorta, either as part of a more extensive dissection (DeBakey I) or in isolation (DeBakey II). Type B (distal or DeBakey III) dissections arise after the take-off of the great vessels. Type A dissections typically have the most serious complications and greatest associated mortality.

Clinical Features. The morbidity and mortality associated with dissections depend on which part of the aorta is involved; the most serious complications occur with dissections between the aortic valve and the distal arch. Accordingly, aortic dissections are generally classified into two types (Fig. 11-22).

- The more common (and dangerous) *proximal* lesions (called *type A dissections*), involving either both the ascending and descending aorta or just the ascending aorta only (types I and II of the DeBakey classification)
- *Distal lesions not involving the ascending part* and usually beginning distal to the subclavian artery (called *type B dissections* or DeBakey type III)

The classic clinical symptoms of aortic dissection are the sudden onset of excruciating pain, usually beginning in the anterior chest, radiating to the back between the scapulae, and moving downward as the dissection progresses; the pain can be confused with that of myocardial infarction.

The most common cause of death is rupture of the dissection into the pericardial, pleural, or peritoneal cavities. Retrograde dissection into the aortic root can also disrupt the aortic valve annulus. Common clinical manifestations include cardiac tamponade and aortic insufficiency. Dissections can also extend into the great arteries of the neck, or into the coronary, renal, mesenteric, or iliac arteries, causing vascular obstruction and ischemic consequences such as myocardial infarction; involvement of spinal arteries can cause transverse myelitis.

In type A dissections, rapid diagnosis and institution of intensive antihypertensive therapy coupled with surgical plication of the aortic intimal tear can save 65% to 85% of patients. However, mortality approaches 70% in those who present with hemorrhage or symptoms related to distal ischemia, and the overall 10-year survival is only 40% to 60%. Most type B dissections can be managed conservatively; patients have a 75% survival rate whether they are treated with surgery or antihypertensive medication only.

KEY CONCEPTS

Aneurysms and Dissections

- Aneurysms are congenital or acquired dilations of the heart or blood vessels that involve the entire thickness of the wall. Complications are related to rupture, thrombosis, and embolization.
- Dissections occur when blood enters the wall of a vessel and separates the various layers. Complications arise due to rupture or obstruction of vessels branching off the aorta.
- Aneurysms and dissections result from structural weakness of the vessel wall caused by loss of smooth muscle cells or insufficient extracellular matrix, which can result from ischemia, genetic defects, or defective matrix remodeling.

Vasculitis

Vasculitis is a general term for vessel wall inflammation. The clinical features of the various vasculitides are protean and largely depend on the vascular bed affected (e.g., CNS vs. heart vs. small bowel). Besides the findings referable to the specific tissues involved, the clinical manifestations typically include constitutional signs and symptoms such as fever, myalgias, arthralgias, and malaise.

Vessels of any type in virtually any organ can be affected, but most vasculitides affect small vessels ranging in size from arterioles to capillaries to venules. There are exceptions, however, and, several of the vasculitides tend to affect only vessels of a particular size or location. Thus, there are entities that primarily affect the aorta and medium-sized arteries, while others principally affect only smaller arterioles. Some 20 primary forms of vasculitis are recognized, and classification schemes attempt (with variable success) to group them according to vessel diameter, role of immune complexes, presence of specific autoantibodies, granuloma formation, organ specificity, and even population demographics (Table 11-3 and Fig. 11-23). As we will see, there is considerable clinical and pathologic overlap among these entities.

The two common pathogenic mechanisms of vasculitis are immune-mediated inflammation and direct invasion of vascular walls by infectious pathogens. *Infections can also indirectly induce a noninfectious vasculitis,* for example, by generating immune complexes or triggering a cross-reactive immune response. In any given patient, it is critical to distinguish between infectious and immunologic mechanisms, because immunosuppressive therapy is appropriate for immune-mediated vasculitis but could very well be counter-productive for infectious vasculitides. Physical and chemical injury, such as from irradiation, mechanical trauma, and toxins, can also cause vasculitis.